Constantinou, C., Payne, N. & Inusa, B. (2015). Assessing the Quality of Life of Children with Sickle Cell Anaemia using Self-, Parent-proxy and Healthcare Professional-proxy Reports. *British Journal of Health Psychology, 20,* 290-304. DOI: 10.1111/bjhp.12099

Assessing the Quality of Life of children with Sickle Cell Anaemia using self-, parent-proxy and healthcare professional-proxy reports

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Abstract

Objectives. The Quality of Life (QoL) of children with Sickle Cell Anaemia (SCA) in the UK has not been examined, and a discrepancy measure based on Gap theory has rarely been used. This study investigated whether 1) child self-reports of QoL using a discrepancy measure (the Generic Children's QoL Measure; GCQ) are lower than those from healthy children, 2) proxy reports from parents and healthcare professionals are lower than child self-reports, and 3) demographic and disease severity indicators are related to QoL.

Design and Methods. An inter-dependent groups cross-sectional design was implemented. Seventy-four children with SCA, their parent and members of their healthcare team completed the GCQ. Demographic and disease severity indicators were recorded. GCQ data from healthy children were obtained from the UK Data Archive.

Results. Contrary to past research, when examining generic discrepancy QoL, children with SCA did not report lower QoL than healthy children, and parent- and healthcare professional proxy reports were not lower than child self-reports. Few of the demographic and disease severity indicators were related to QoL.

Conclusions. Proxy reports may be used to gain a more complete picture of QoL but should not be a substitute for self-reports. The explanation for the relatively high levels of QoL reported is not clear, but children with SCA may have realistic expectations about their ideal self, place greater emphasis on aspects other than health in shaping their QoL, and define achievements within the limits of their illness. Future research should focus on psychological factors in explaining QoL.

Keywords: paediatric; sickle cell anaemia; quality of life; gap theory; disease severity.

Introduction

Homozygous SS (HbSS) otherwise referred to as sickle cell anaemia (SCA) is the most prevalent and severe form of sickle cell disease (SCD). It has potentially life-threatening complications such as anaemia and infections and can cause stroke and multi-organ damage. These complications along with pain can lead to frequent hospital admissions and days missed from school. As a result, it is likely to have physical, psychological, and social consequences and thus may impair quality of life (QoL; Peterson, 2009). Using health-related QoL measures such as the Paediatric Quality of Life Inventory (PedsQL) and the Child Health Questionnaire (CHQ), studies have found that children with SCD have an impaired health-related QoL compared with healthy children (Palermo, Schwartz, Drotar, & McGowan, 2002; Panepinto & Bonner, 2012; Panepinto, O'Mahar, DeBaun, Loberiza, & Scott, 2005; Panepinto, Pajewski, Foerster, Sabnis, & Hoffmann, 2009).

Quality of life is most commonly obtained from child self-report or proxy report, which is most often sought from parents (Varni, Burwinkle, & Lane, 2005). Proxy reports are useful when children are unable to complete measures themselves due to young age, physical illness, emotional distress, or cognitive impairment (Daly, Kral, & Tarazi, 2011). Such reports may be most useful if they are relatively accurate when compared to child self-reports. However, they may provide a more complete picture of child QoL even if differences exist (Upton, Lawford, & Eiser, 2008). In a review by Eiser and Morse (2001), higher agreement between child and parent-proxy reports of QoL was found for observable physical aspects of QoL compared with emotional or social aspects. In a further review, generally good agreement was found, but parents of children with health conditions tended to rate their child's QoL lower than the child's self-report (Upton et al., 2008). The same was found in research on children with SCD (Dale, Cochran, Roy, Jernigan, & Buchanan, 2011; Panepinto et al., 2005), although sometimes differences were only obtained on the physical and not

psychosocial domain of QoL (Barakat, Patterson, Daniel, & Dampier, 2008).

Parents may be the preferred choice of proxy rater as they observe the child engaging in a wide range of activities and may have better access to his/her thoughts and feelings. Health care professionals (e.g., physicians and nurses) see the child in restricted situations, so may find it more difficult to accurately estimate a child's QoL. However parent ratings may be biased by their caregiving function and their own anxieties and concerns. Therefore, as both parents and health care professionals play a central role in making treatment decisions, it is important to understand any discrepancies between child and proxy reports of QoL as these may influence a child's care (Dampier, Ely, Brodecki, & O'Neal, 2002; Varni et al., 2005). In research on children with psychiatric disorders, amoderate correlation between reports from children and parents was found as well as a small correlation between reports from children and their clinician (Bastiaansen, Koot, Ferdinand, & Verhulst, 2004). Health care professional reports have not been examined in the paediatric SCD population, but evidence suggests that children with SCD tend to perceive less symptom severity compared with physicians, and objective measures of disease severity may in part account for this discrepancy (Connelly et al., 2005).

Child self-reports and adult-proxy reports of QoL may be better understood by taking into account demographic and disease severity indicators (Upton et al., 2008). Most research in this area has focused on parent-proxy reports and findings vary. For example, older and female children, a greater number of chronic transfusions, and number of complications were associated with poorer physical, but not psychosocial parent-proxy QoL measured by the CHQ (Palermo et al., 2002). Overall disease severity, being an older child, having lower family income, and co-morbidities were associated with poorer physical and psychosocial parent-proxy QoL, but only overall disease severity was associated with child self-reported QoL using the PedsQL (Panepinto et al., 2009). Days missed from school were also found to

be related to child and parent-proxy reports (Dampier et al., 2010). There has been no research into the variables that may be related to health care professional-proxy reports in the paediatric SCD population, although the degree of disease severity may be a contributing factor (Connelly et al., 2005).

The majority of previous research on QoL uses measures that assess individuals' perceptions of their current state. However, QoL is defined by the World Health Organization (WHO) as an individual's current state in comparison with their expectations (WHO-QOL Group, 1995). Therefore, most measures of QoL, such as the CHQ and PedsQL, fail to adopt the WHO's definition and do not assess the discrepancy between an individual's perceived current state and their expectations (Quittner, Cruz, Modi, & Marciel, 2009). However, Gap theory involves measuring this discrepancy (Calman, 1984). Such an approach is important because it may help explain how individuals with a chronic illness or disability do not necessarily rate themselves to have poor QoL (Albrecht & Devlieger, 1999). Furthermore, it has implications for intervention, as it suggests that QoL may be improved by helping people adjust their expectations or by helping them develop and grow in other ways.

Previous research using this approach has mainly focused on adults. Research on adjustment to cancer has found that a high discrepancy between actual-and ideal-self leads to poor adaptation and psychological distress (Heidrich, Forsthoff, &Ward, 1994). As time passes, people may lower their expectations and thereby reduce discrepancy. For example, Heidrich and Ward (1992) found that women with cancer had lower actual-and ideal-self ratings than women without cancer but were no different in terms of discrepancies between the two and adjustment.

Welham, Haire, Mercer, and Stedman (2001) developed the QoL-GAP instrument and found that in adults with severe mental illness, discrepancy QoL scores were consistently more associated with life satisfaction compared with current QoL scores alone. Another

discrepancy measure, the QoL Assessment Schedule has been used in adults with epilepsy (Selai, Elstner, & Trimble, 2000) and dementia (Selai, Trimble, Rossor, & Harvey, 2000).

When comparing children with asthma to healthy children, Eiser, Vance, and Seamark (2000) found that they had higher discrepancy scores on the Exqol. Significant correlations were also found between scores on this discrepancy measure and an asthma-specific QoL scale and self-ratings of asthma severity. Collier, MacKinlay, and Phillips (2000) developed the Generic Children's Quality of Life Measure (GCQ). Using this measure, Heath et al. (2011) found that children with Chronic Kidney Disease (CKD) actually had better QoL scores than healthy children.

The GCQ was designed to allow comparison between healthy children and those with a chronic illness (Collier & MacKinlay, 2008; Collier et al., 2000; Heath et al. , 2011). It was developed using constructs provided by children when asked about QoL and designed to be more child-friendly than other measures of paediatric health-related QoL in terms of its lay-out and use of a story format. Furthermore, whereas most research on paediatric SCD uses health-related QoL measures, the GCQ covers general areas such as education, family, and peer relationships. The authors argue that measures of health-related QoL include an assessment of health status, and so it is unsurprising when using such measures that children with chronic illnesses are seen to have a reduced QoL compared with healthy children. Furthermore, research on adults with cancer (e.g., Heidrich et al., 1994) or a chronic illness or disability (Albrecht & Devlieger, 1999) suggests that on a social and emotional level, adults may adapt well to illness and thus may not experience a reduced QoL. The same may apply to children. Thus, Collier et al. (2000) and Heath et al. (2011) suggest that it is more appropriate to use a generic psychosocial QoL measure based on Gap theory. However, research has not examined a Gap theory approach applied to SCD.

The present study

This study focused on children with the most severe form of SCD – homozygous SS (SCA). A recent review found no published research that had used health-related QoL measures to assess QoL in paediatric SCD in the United Kingdom (Panepinto & Bonner, 2012), so the present study appears to be the first in the United Kingdom. Furthermore, most research on paediatric QoL uses the PedsQL or CHQ which fail to adopt the WHO's discrepancy definition of QoL based on Gap theory. Thus, the present study is also the first to apply a discrepancy measure (the GCQ) in this population. For this reason, it is important to compare GCQ scores obtained in the present study to those obtained from healthy children. As children may sometimes be unable to complete measures themselves, it is also important to examine both child self-reports and relevant proxy reports of QoL and to consider what may determine QoL reports and any lack of concordance between reports from different informants. Therefore, the research questions addressed in the present study are as follows:

1. Do SCA children have lower self-reported GCQ discrepancy scores compared with a matched sample of healthy children?

2. Are parent-proxy and health care professional-proxy reported GCQ scores lower than child self-reports?

    a. If differences in GCQ discrepancy scores exist, which demographic and disease severity indicator variables are associated with these differences?

3. Which demographic and disease severity indicator variables are related to child self reported, parent-proxy and health care professional-proxy reported GCQ discrepancy scores?

Methods

Participants

A total of 205 children with SCA and their parents currently attending the haematology out patient clinic at a London hospital were contacted by post and invited to

participate in the study. Children were identified as HbSS phenotype from the clinic register of patients. A power calculation using G* Power3 (Faul, Erdfelder, Lang, & Buchner, 2007) showed that to examine the relationship between child and proxy measures of QoL, a sample size of 75 would be sufficient to detect a small to medium effect with 95% power and a 5% significance level.

Seventy-four children with SCA aged 6– 16, and their parents volunteered to participate (response rate 37%). Sample demographics and disease severity indicators are shown in Table 1. Additionally, four paediatric clinical nurse specialists who have known the children and their families for many years, and see and/or speak on the phone to the child at least quarterly, were invited to participate by providing health care professional-proxy QoL reports.

As there were no available GCQ data for healthy children aged over 14, self-reported data from 65 healthy children were selected from the UK Data Archive (UK Data Archive, 2008) for comparison with the 65 children aged 6– 14 in the SCA sample in the present study. The Archive included GCQ data from 720 healthy children (347 males). The sample was gender and age matched by implementing a quota sampling technique.

Materials

The GCQ. There are two identical gender-specific versions of this measure, which is presented as a child-friendly booklet. The first section includes 25 questions that ask the child who is most like them (their perceived-or current-self). For example, ‘how often they have enough friends’, ‘how often they worry about things’, ‘how often they are good at sport’, and ‘how often they feel different from other children’. The second section includes the same 25 questions that ask the child who they would most like to be like (their preferred or ideal-self). All of the items are scored on a 5-point Likert scale (‘always’, ‘often’, ‘sometimes’, ‘hardly ever’, and ‘never’).

Scores are calculated by measuring the discrepancy between a participants' perceived current-self score and their preferred ideal-self score. The discrepancy score ranges from 0 to 100 and is calculated by subtracting the perceived current-self score from the preferred ideal-self score for each item and then subtracting the sum of these discrepancy scores from 100. A higher score demonstrates higher QoL. Norm discrepancy QoL scores from healthy children aged 6– 14 are 73.7 (SD = 9.9) for boys and 74.7 (SD = 10.0) for girls (Collier et al. , 2000).

In the present study, child self-reports, parent-proxy reports, and health care professional-proxy reports of perceived current-self, preferred-self, and the discrepancy score all demonstrated good reliability (a ranged from .75 to .94). Collier et al. (2000) and Heath et al. (2011) found reliabilities ranged from .74 to .78, and discrepancy scores correlated with a single item measure of QoL ($r = .50$, $p < .001$), supporting construct validity.

Demographic indicators. Demographic indicators included the child's gender and age, and the parent's gender, marital status (married/cohabiting or single), and employment status (employed or unemployed).

SCA severity measures. Measures of disease severity (three obtained from parents' reports and three obtained from hospital records) were chosen based on those used in previous research (e.g., Palermo et al., 2002). They were not combined to form a total measure but were treated separately. The severity measures completed by parents included the total number of crises a child had experienced in the last 12 months, such as frequent painful episodes requiring physician attention, severe anaemia, or infections; the number of hospital admissions and accident and emergency (A&E) visits; and the number of days a child had missed from school due to SCA in the last 12 months.

Measures collected from the children's hospital records included the number of hospital admissions and A&E visits, and indicators of stroke risk measurements. Stroke is a good measure of disease severity because it is one of the most devastating complications of

SCD (Ballas et al., 2010). The risk of developing stroke is determined by the degree of vascular stenosis, which is reliably estimated using transcranial Doppler scan (TCD) velocity (i.e., velocity of blood flow in the cerebral vessels). TCD velocity is now accepted as a risk factor that warrants consideration for bone marrow transplantation. Thus, it is used routinely to assess the risk of stroke in SCA. It involves two main arteries: internal carotid arteries (left and right ICA) and middle cerebral arteries (left and right MCA). TCD is categorized as normal (< 170 cm/s), conditional (170– 199 cm/s) with a moderate risk of stroke, and abnormal readings (> 199 cm/s) with a high risk of stroke (Brambilla, Miller, & Adams, 2007). The brain structure was also verified with magnetic resonance imaging (MRI), and images were categorized as either normal or abnormal based on whether or not there was evidence of infarction or cerebrovascular abnormalities such as lesions.

Procedure

After ethical approval was obtained for this study, the child and their parent were sent age appropriate information sheets to their home address 1 week before data collection commenced. Children and their parent were then approached during their outpatient clinical appointment. The researcher read through age-appropriate information sheets with all of the participants, answering any questions to ensure they understood before deciding whether to participate. The parent and child were asked to give consent for their own participation. The parent was also asked to provide consent for their child's participation and was given a demographic and disease severity information sheet to complete.

Children, their parent and a member of the health care team were asked to independently complete the GCQ. If the child had understood the practice question, they would then fill in the GCQ with the researcher in a quiet area in the waiting room away from their parent. Parents and members of the health care team were asked to complete the GCQ as if they were the child. Parents independently completed the measure in the waiting room

away from their child and staff.Members of the health care team were asked to complete the measure at a convenient time, on the same day as the child's outpatient clinical appointment.

Analysis

IBM SPSS Statistics for Windows, Version 21.0 (IBM Corp., Armonk, NY, USA) was used to analyse the data. To address research question 1 (do SCA children have a lower QoL compared with a matched sample of healthy children using child self-reported QoL discrepancy scores?), an independent samples **t**-test was used. To address research question 2 (are parent-proxy reports and health care professional-proxy reports of QoL on the GCQ lower than child self-reports?), intraclass correlation coefficients (ICC) were calculated and paired-samples **t**-tests were conducted to examine child–parent and child–health care professional concordance on reports of perceived current-self and preferred ideal-self, and on the discrepancy scores (following e.g., Cremeens, Eiser, & Blades, 2006). Where significant child–parent and child–health care professional differences are found in discrepancy scores, repeated-measures ANCOVAs may be used to ascertain which disease severity and demographic indicators are associated with any differences (following, for example, Britto **et al.**, 2004). To address research question three (are disease severity and demographic indicators related to child self-, parent-proxy, and health care professional-proxy reported QoL discrepancy scores?), preliminary univariate analyses were first performed. Pearson's correlation analyses were conducted for stroke risk measures and age, and independent samples **t**-tests for dichotomous variables (i.e., child's gender, parent's marital status, gender and whether employed, and MRI classification). As the remaining disease severity indicators were not normally distributed, categorical variables were created, each with three levels, and one-way ANOVAs, followed by Tukey's HSD post-hoc tests, were performed. This was done for total number of SCA crises (0, 1, and ≥2), days missed from school (0, ≤2, and >2 weeks), and hospital admissions and A&E visits from parents' reports and from hospital records (0, 1,

and ≥2). These were followed by three multiple regression analyses to examine the relationships between child, parent-proxy, and health care professional-proxy QoL discrepancy scores and the variables that were statistically significant in the univariate analyses. Any categorical variables entered in the regressions were dummy-coded.

Results

Do SCA children have lower self-reported QoL discrepancy scores compared with a matched sample of healthy children?

Child self-reported QoL discrepancy scores from children with SCA (M = 73.34, SD = 9.80) were compared with the matched sample of healthy children (M = 74.47, SD = 9.92), and there was no significant difference, $t(67) = -1.11$, $p = .27$. Therefore, children with SCA do not appear to have lower QoL compared with healthy children.

Insert Tables 1 and 2 about here

Are parent-proxy and health care professional-proxy reports of QoL lower than child self-reports?

Child–parent and child–health care professional concordance in QoL scores was explored as shown in Table 2. Parent-proxy-perceived current-self scores were significantly lower than child self-reports and the ICC was significant. This suggests that the difference between scores is consistent across individuals. There was also a smaller significant ICC between child- and health care professional-perceived current-self scores. Health care professional-proxy-preferred ideal-self scores were significantly higher than child self-reports and the ICC was not significant. This suggests that the difference was not consistent across individuals. Finally, there were no significant differences between child and parent-proxy, and child and health care professional-proxy discrepancy scores, but the ICCs were significant. As

the ICC between child and parent scores was very small, this suggests that health care professionals may be more accurate in gauging a child's discrepancy QoL than parents. As there were no significant differences between child self and proxy reports, further analyses to ascertain demographic and disease severity indicators associated with differences were not conducted.

Which demographic and disease severity indicators are related to child self-reported, parent-proxy, and health care professional-proxy reported QoL discrepancy scores?

Relationships between demographic and disease severity indicators and child self-reported, parent-proxy, and health care professional-proxy reported QoL discrepancy scores are shown in Table 3. Children who had not missed any days of school in the previous 12 months (M = 79.81, SD = 6.02) self-reported higher QoL compared with children who had missed up to 2 weeks (M = 71.52, SD = 10.17) and who had missed more than 2 weeks (M = 73.12, SD = 10.41).

Health care professional-proxy reports of QoL were higher for children who had missed zero days of school in the previous 12 months (M = 78.38, SD = 9.08) or up to 2 weeks (M = 72.00, SD = 12.13) compared with children who had missed more than 2 weeks (M = 64.64, SD = 12.22). Also, health care professional-proxy reports of QoL were higher for children who had experienced zero (M = 75.91, SD = 10.76) or one crisis (M = 72.22, SD = 11.25) in the previous 12 months compared with children who had experienced two or more crises (M = 65.04, SD = 13.32).

Finally, parents who were single (M = 67.66, SD = 12.97) provided lower proxy QoL scores compared with those who were married or co-habiting (M = 75.61, SD = 10.22). Also, higher parent-proxy scores were correlated with higher left ICA and higher left MCA measurements. This is unexpected as one might expect higher parent-proxy scores in children with less severe symptoms as indicated by stroke risk measurements. Further Pearson's

correlation analyses (not shown for reasons of brevity) indicated that stroke risk measurements were not related to any other measures of disease severity used in the current study (all $p > .05$), which may suggest that these stroke risk measures are not a good reflection of the degree of disease severity experienced by families.

Regression analyses were run including variables significant in the univariate analyses. A regression was not run for child self-reports because there was only one statistically significant finding in the univariate analyses. The regression models explained 15% and 16% of the variance in parent-proxy and health care professional-proxy reported QoL discrepancy scores, respectively, as shown in Table 4. Only marital status was statistically significant in the first regression, while only the dummy variable zero days missed from school due to SCA was statistically significant in the second regression.

Insert Tables 3 and 4 about here

Discussion

Do SCA children have lower self-reported QoL discrepancy scores compared to a matched sample of healthy children?

Children with SCA were not found to have lower QoL discrepancy scores. This is contrary to previous research, which has found that children with SCA have a lower health-related QoL than healthy children, particularly on the physical domain (Panepinto & Bonner, 2012). According to Collier et al. (2000) and Heath et al. (2011), it may not be surprising that children with chronic illnesses are seen to have a reduced QoL when past research uses measures that include an assessment of health status. They argue that as children with chronic illnesses, such as SCA, generally lead similar lives to, and interact on a daily basis with their healthy peers, they are more likely to assess QoL in relation to these healthy children. Indeed,

past research using the GCQ has found that children with CKD actually have higher discrepancy QoL scores than healthy children (Heath et al. , 2011). Similar findings have been observed in children with asthma using a different measure based on Gap theory (Eiser et al. , 2000). Research in adults with cancer (e.g., Heidrich et al. 1994) or chronic illness or disability (Albrecht & Devlieger, 1999) suggests that adults may socially and emotionally adapt well to illness, most likely by lowering their expectations to reduce discrepancy between their actual- and ideal-self; thus, they may not experience a reduced QoL. In the present study, children with SCA did not report any impairment in generic QoL, although as children are born with SCA, the adaptation mechanism suggested above may not apply. Instead, children's personal experience with SCA from the beginning of life in defining the self and their view of the world may mean they have realistic expectations about their ideal-self (Dickinson et al. , 2007). Furthermore, other aspects of their experiences rather than their health status may play a greater role in shaping their QoL. Additionally, their concept of success may be a sense of achievement within the limits of their illness (Albrecht&Devlieger, 1999). Thus, they may experience a good QoL on a social and emotional level, which is what was assessed in the present study using a generic QoL measure.

Are parent-proxy and health care professional-proxy reports of QoL lower than child self-reports?

When using a discrepancy measure of QoL, parent- and health care professional-proxy scores were not lower than the child self-reports, which is contrary to much previous research (Panepinto et al., 2005; Upton et al., 2008). This contrast may in part be explained by the fact that past research assessed health-related QoL. Indeed, previous research has tended to find that parent-proxy reports are more often lower than child self-reports on the physical domain of health-related QoL (which is specific to health-related QoL) than on the psychosocial domain (which is more similar to the generic QoL measured in the present study; Barakat et

al., 2008). Furthermore, past research has generally examined perceived current QoL and not the discrepancy between perceived current- and preferred ideal-self. Indeed, in the present study, parent-proxy reports of perceived current QoL were lower than child self-reports, suggesting that using discrepancy scores makes a difference to child– parent concordance.

Health care professional-proxy discrepancy scores were also not lower than child self-report scores. The significant ICC between these scores, compared with the smaller ICC between child and parent scores, suggests that health care professionals may be slightly more accurate in gauging a child's discrepancy QoL. This is unexpected because it was anticipated that this would be more difficult for health care professionals given the restricted situations in which they see the child. However, it was also found that health care professionals report on average higher ideal QoL scores than child self-reports. Thus, it appears to be the case that children, and also parents to some extent, have adapted their expectations (reflected in their ideal QoL scores), whereas health care professionals have generally high expectations about what is ideal. The focus in past research on perceived current QoL may therefore be misleading as this does not take into account differences in expectations about ideal QoL that are part of a discrepancy measure.

Which demographic and disease severity indicators are related to child self-reported, parent-proxy, and health care professional-proxy reported QoL discrepancy scores?

The present study found few significant predictors of QoL. Child self-reported discrepancy QoL was higher for children who had not missed any days of school. Furthermore, in multivariate analyses, married or cohabiting parents reported a higher parent-proxy discrepancy QoL compared with those who were single, and health care professional-proxy-reported discrepancy QoL was higher for children who had not missed any days of school, supporting previous research (Dampier et al. , 2010). In contrast, previous research has found that disease severity is an important correlate of parent- proxy-reported health-

related QoL (Palermo et al., 2002; Panepinto et al., 2005, 2009), and it might be expected that this would be even more likely to explain health care professional-proxy reports (Connelly et al. , 2005). Once again, the reason this was not more important in the present study is likely to be due to the focus on generic QoL. Indeed, past research has found that disease severity is more likely to be associated with the physical than the psychosocial domain of health-related QoL (Palermo et al., 2002; Panepinto et al., 2005). Also, the present study may not have incorporated the most relevant disease severity measures. Additionally, treatments such as chronic transfusion and hydroxyurea that may have helped explain QoL were not included in the present study. Furthermore, as previous research has found differences in QoL between children with mild and severe SCD (Panepinto et al., 2005, 2009), it may be that the level of disease severity in the sample in the present study was relatively mild (e.g., only 14% of the sample had abnormal MRI scans). Nevertheless, it is reassuring that disease severity, as measured in the present study, does not appear to impair generic QoL, but it is not clear what is important in explaining it. Particularly in the case of child and parent-proxy-reported QoL, psychological variables may be key. It has been suggested that efforts to improve QoL in SCD should incorporate a focus on adolescent psychological functioning (namely reduction of anxiety and depression) and disease-related parenting stress (Barakat et al., 2008).

Implications and directions for future research

As children's sense of self from the start of life incorporates SCA, they may have realistic expectations about their ideal-self, leading to little discrepancy between their actual- and ideal-self (Dickinson et al., 2007). However, as health is one of the many components of QoL, focusing the concept of QoL on health-related issues may reduce SCA children to patients with poor health and diminished function (Albrecht & Devlieger, 1999). In the traditional model of health care, interventions focus on restoring health, but for children with SCA, identifying whether and where discrepancies between actual- and ideal-self exist

(beyond norms for healthy children) and helping them adjust their expectations may be required (Carr, Gibson, & Robinson, 2001; Eiser et al., 2000). The idea is not to lower expectations so that children tolerate significant levels of disease and disability but to help them have more realistic expectations. Carr et al. (2001, p. 1242) suggest that 'reflecting on aspects of reality is part of the process of empowerment and provides the force that allows people to take action to change that reality'. Children with SCA may be likely to have lower QoL on the physical domain of health-related QoL (as found in previous research, for example Panepinto & Bonner, 2012), and significantly reducing discrepancies to within the norms for healthy children may be unrealistic. However, focusing on generic QoL, which is somewhat similar to the psychosocial domain of health-related QoL, provides greater possibilities for improving QoL. Interventions aimed at improving sense of control and social relationships and networks could be helpful. Research should focus on developing appropriate interventions to reduce discrepancies and thus improve QoL.

A generic QoL measure such as the GCQ would be especially appropriate for use by health and clinical psychologists when working with children with SCA. It would be interesting to also examine psychologist-proxy reports of QoL to see how these differ from child self- and other proxy reports. Psychologists could also use the GCQ to assess the impact of interventions aimed at improving QoL. Additionally, the GCQ could be used to assess the impact of different medical treatments. It would be important to know whether it is sensitive to changes in QoL over time during different stages of SCA treatment.

Limitations

A number of limitations of the present study have already been discussed, such as not including measures of treatment and other measures of disease severity and relatively low levels of disease severity in the sample. The use of a generic uni-dimensional QoL measure, which is not directly comparable to health-related QoL measures used in past research and

does not include multiple dimensions, may also be considered a limitation, although the benefits of using such a measure have been discussed. Additionally, the study had a limited sample size from one hospital, with relatively few male child and male parent participants, and few older adolescents, thus limiting the generalizability of the findings. Furthermore, parent reported measures of disease severity may not be accurate, although parent reports of number of hospital admissions and A&E visits were similar to reports from hospital records. Finally, the study adopted a cross-sectional design, and so changes in QoL over time were not evaluated and causal relationships cannot be established.

Conclusions

The present study appears to be the first to measure QoL in paediatric SCA in the United Kingdom and the first to use multi-informant reports to help provide a more complete understanding of QoL in paediatric SCA. Furthermore, it is also the first study to use a discrepancy measure (the GCQ) based on theory (i.e., Gap theory) in the paediatric SCA population. This provides a more appropriate way of operationalizing QoL, which is also in line with the WHO's definition of QoL. The GCQ may also be a good measure as it is particularly child-friendly.

When using a discrepancy measure, parent- and health care professional-proxy reports were not lower than child self-reports, and there was evidence that health care professionals may be slightly more accurate in gauging a child's discrepancy QoL than parents. However, if the aim of using proxy reports is as a substitute for the child's self-report, proxy reports are unlikely to be sufficiently reliable, as the ICCs between child self and proxy reports were not large. Despite this, incorporating proxy measures may be helpful in gaining a better understanding of a child's QoL.

This study has also shown that when using a discrepancy measure, children with SCA do not report a lower QoL compared with healthy children. It is not clear from the present

study what explains their relatively high levels of QoL. However, the premise for using a generic discrepancy measure of QoL may lead us to suggest that children with SCA might have realistic expectations about their ideal-self, place greater emphasis on aspects other than health in shaping their QoL, and define achievements within the limits of their illness.

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Table 1

Descriptive statistics for demographic and disease severity indicators

	M (*SD*) or % n
Child age	10.61 (3.10)
Child gender	65% female, 35% male
Parent age	41.31 (6.80)
Parent gender	76% female, 24% male
Parent marital status	49% married/cohabiting, 51% single/divorced
Parent employment status	65% employed, 35% unemployed
Number of crises in 12 months	30% no crises, 36% 1 crisis, 34% ≥2 crises
Days missed from school in 12 months	22% no days, 45% ≤ 2 weeks, 33% >2 weeks
Hospital and A&E admissions in 12 months (parent report)	53% no admissions, 24% 1 admission, 23% ≥2
Hospital and A&E admissions in 12 months (hospital records)	45% no admissions, 27% 1 admission, 28% ≥2
Right ICA	93.26cm/sec (21.69)
Left ICA	98.93cm/sec (23.44)
Right MCA	118.07cm/sec (27.18)
Left MCA	120.04cm/sec (25.94)
TCD classifications	90% normal, 7% conditional, 3% abnormal
MRI	86% normal, 14% abnormal

Table 2

Pearson's correlations and t-tests examining concordance in QoL scores between child and parent-proxy reports and child and healthcare professional-proxy reports

	M (*SD*) participant reports of QoL			Child-parent		Child-healthcare	
	Child	Parent	Healthcare	r	t	r	t
Perceived current	88.69 (9.96)	85.51 (9.45)	87.64 (14.03)	.45***	2.88**	.20*	.58
Preferred	108.59 (10.48)	108.43 (11.86)	113.76 (5.70)	-.03	.09	-.17	-3.55**
Discrepancy	73.12 (10.41)	71.53 (12.30)	70.89 (12.52)	.19*	.95	.36***	1.48

Note. * p < .05, ** *p* < .01. *** p < .001.

Table 3

T-tests, ANOVAs and pearson's correlations examining relationships between demographic and disease severity indicators and child-, parent-proxy and healthcare professional-proxy reports of QoL

	Child-self reported QoL	Parent proxy-reported QoL	Healthcare proxy-reported QoL
T-tests	*t*	*t*	*t*
Parent's gender	1.79	-.01	.62
Parent's marital status (single, married/cohabiting)	.33	2.92**	-1.02
Parent's employment status (employed, unemployed)	1.06	1.31	-.60
Child's gender	1.89	-1.02	1.52
MRI scan (normal, abnormal)	.40	.34	1.96
ANOVAs	F	F	F
Number of crises	1.69	1.65	5.18**
Days missed from school	4.66*	.74	7.13**
Hospital and A&E admissions (parent report)	1.99	.73	1.64
Hospital and A&E admissions (hospital report)	2.26	.22	1.16
Correlations	r	r	r
Child's age	.02	-.05	.06

Right ICA	.07	.13	-.04
Left ICA	.07	.26*	-.03
Right MCA	.01	.19	-.17
Left MCA	.13	.29*	-.04

Note: * $p < .05$, ** $p < .01$.

Table 4

Two standard multiple linear regression analyses examining parent-proxy and healthcare professional-proxy reports of QoL

Participant reports	Predictor	*β*	*t*	Partial correlations	Contribution of variable to total % of explained variance
Parents[a]	Marital status (single, married /cohabiting)	-.28	-2.54*	-.29	8.41
	Left ICA	.13	1.09	.13	1.69
	Left MCA	.20	1.68	.20	4.00
Healthcare professionals[b]	Zero crises (dummy)	.19	1.25	.15	2.25
	One crisis (dummy)	.22	1.74	.21	4.41
	Zero days missed (dummy)	.38	2.51*	.29	8.41
	≤ Two weeks missed (dummy)	.24	1.89	.22	4.84

Note. [a] $R^2 = .15$, $R = .43$, $F(3,70) = 5.22$, $p = .003$. [b] $R^2 = .16$, $R = .45$, $F(4,69) = 4.44$, $p = .003$. * $p < .05$